

leukocaria

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➤ A 11-month infant brought by his mother to pediatric ophthalmology clinic , after the mother notices his eyes appeared white in a photograph.

➤ **So, what is the most likely diagnosis ?**



leukocoria

- **Leukocoria** (or **white pupillary reflex**) is an abnormal white reflection from the eye.
- Leukocoria is a medical sign for a number of several conditions.



DIFFERENTIAL DIAGNOSIS OF LEUKOCORIA

- Congenital Cataract
- Retinoblastoma
- **PHPV**
- **Toxocariasis**
- **Coat's disease**
- **ROP**
- **Coloboma of optic disc & retina**
- Retinal detachment
- Norrie's disease



Clinical work-up for child with leukocoria

➤ the differential diagnosis can be narrowed through :

- **History :**

- age of presentation -----

- at birth PHPV , cong. cataract

- 1-3 Y..... RB , cong.cataract

- Pre school age toxocariases , Coats

- birth history

- pre-mature , low birth wight ROP

- **Ocular examination** : EUA, fundus ex.

- **Investigation :**

- B-scan

- CT-scan, MRI

**Persistent hyperplastic
primary vitreous**

Persistent hyperplastic primary vitreous

- It's a congenital anomaly of the eye that **results from** failure of embryological primary vitreous and hyaloid vasculature to regress.

- It is **characterized** by persistence of various portions of the primary vitreous , associated with microphthalmia, cataract and glaucoma.

PHPV is usually divided into **three** types:

1- Anterior PHPV:

- occurs when the remnant vascular stalk is seen attached to the back of the lens but not extended back to the optic nerve.
- This form is typically associated with cataract, glaucoma and retrolenticular membrane.



2- Posterior PHPV:

- The remnant vascular stalk is seen arising off the optic nerve but not reaching the lens, and usually not causing cataract.
- Posterior PHPV may be associated with abnormal development of the retina, optic nerve and macula, vitreal membranes.



3- A combination of anterior and posterior PHPV:

- Is the most commonly seen type.

(a band extending from retina to posterior lens capsule).



- COLOR DOPPLER SHOWING ARTERIALIZED BLOOD FLOW IN THE ECHOGENIC BAND



Treatment

- **Anterior form** is most often treated with observation, lensectomy, I/A with ant. vitrectomy and glaucoma management, whether medical or surgical.
- **Posterior form** is usually associated with a poor visual outcome regardless of intervention due to retinal and optic nerve abnormalities.
- **combined type** is also typically associated with a poor outcome due to the high prevalence of posterior segment abnormalities.

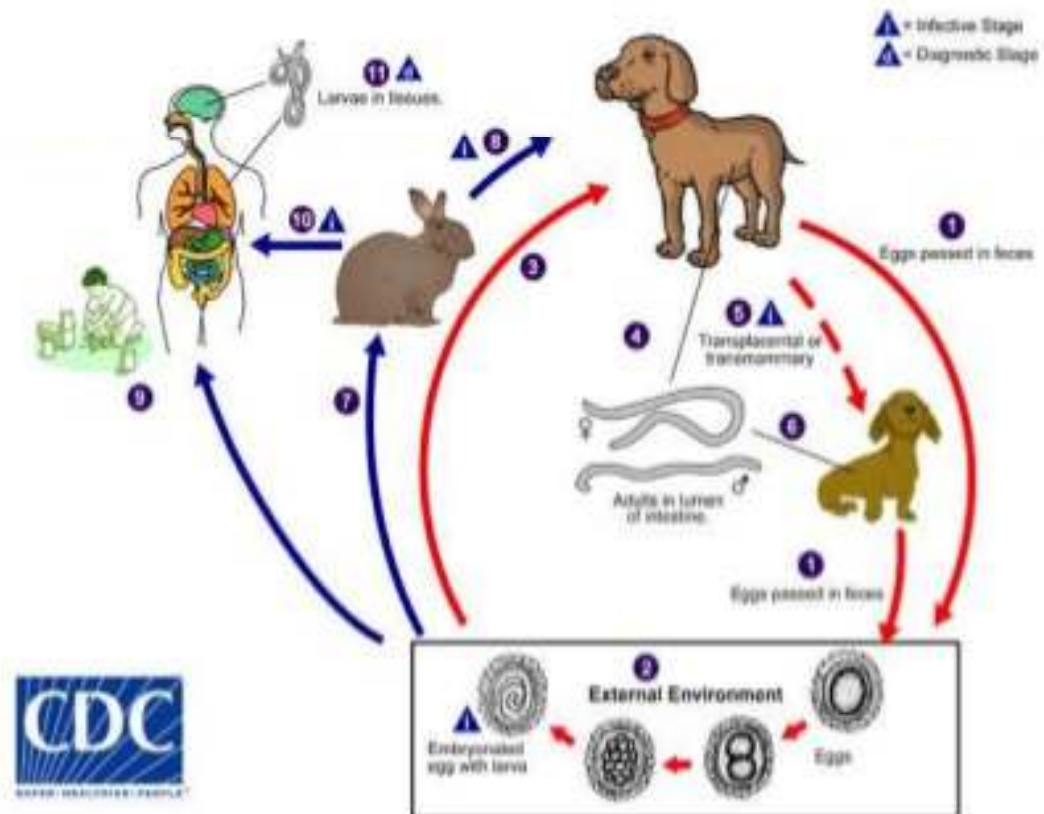
Ocular Toxocariasis

- **Ocular toxocariasis** is an uncommon disease that affects mostly children and young adults, resulting in significant vision loss.

a-Toxocara canis (a common parasite of dogs)

b-Toxocara cati (a common parasite of cats).

- Humans acquire the infection as accidental hosts by ingesting soil contaminated with *Toxocara* eggs ingestion of contaminated food, or the oral-fecal route.



Clinical presentation of Ocular Toxocariasis

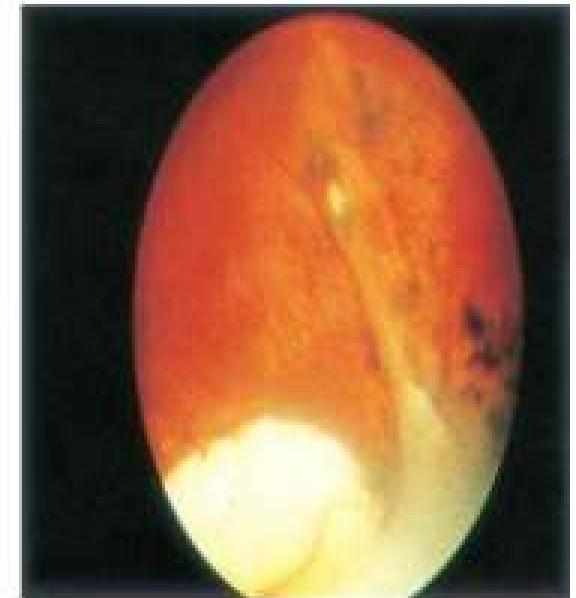
- Chronic endophthalmitis (2-9 years)



(a)

(b)

- Posterior pole granuloma (6-14 years)



- Peripheral granuloma (6-40 years)

TREATMENT

- Medical therapy
 - 5-day course of albendazole (10 mg/kg/day)
 - Consider systemic or periocular corticosteroids to suppress the immune response .
- Surgical therapy (If inflammation persists)
 - Pars plana vitrectomy
 - Perfluorocarbon liquids injection
 - Indicated to facilitate removal of epiretinal membranes (ERMs) and the posterior hyaloid in cases of tractional retinal detachment.
 - Cryotherapy
 - Applied directly at the areas of exudation at the pars plana with a double freeze-thaw technique.
 - Endolaser
 - Indicated for treatment of ocular granulomas

COATS' DISEASE

- **Coats' disease** is caused by(telangiectasia)a problem of blood vessels inside the eye that provide blood and oxygen to the retina.
- In Coats' disease, the blood vessels are dilated, abnormally twisted and leaky.
- This prevents the normal flow of blood, and allows fluid to leak out of the blood vessels (exudate) builds up, it can cause a detachment of the retina and loss of vision.

Coats' Disease

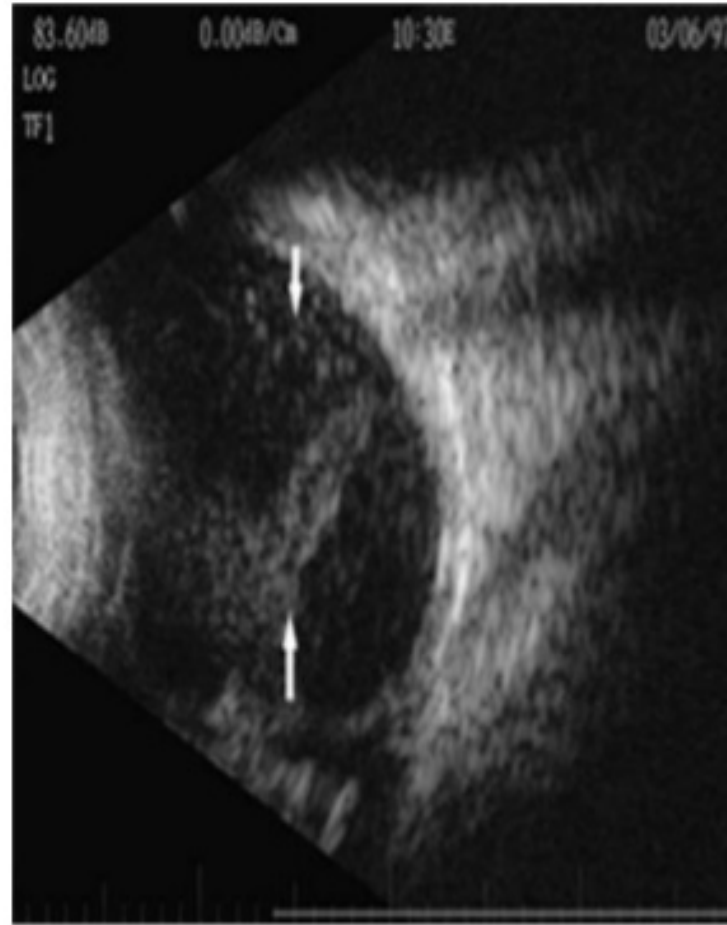




Fig. 1 Retinal detachment visible through pupil in patient with Coats Disease.



- A- A young child Leukocoria (coats' disease). Only visible with a flash camera



- **B SCAN** : AN OPEN FUNNEL RETINAL DETACHMENT WITH DENSE RETRO-RETINAL EXUDATES.
- **CT scan** : of a patient with Coats' disease, showing total exudative retinal detachment in the right eye.

stages of Coats' disease :

- **Stage I:** There is presence of telangiectasia (abnormal blood vessels) only.
- **Stage II:** both telangiectasia as well as exudates.
- **Stage III:** Quite an advanced stage of Coats disease and is characterized by subtotal retinal detachment.
- **Stage IV:** Severely advanced and there is total detachment of the retina.
- **Stage V:** Completely advanced and the affected individual loses vision completely with secondary glaucoma and painful eye.



Treatment

- For **mild form** of Coats Disease , Laser photocoagulation with cryotherapy may be useful for treatment.
- **Advanced stage** : vitreoretinal surgery.
- **If severe pain in the eye**, enucleation is required as a method of treatment.

Retinopathy of prematurity (ROP)

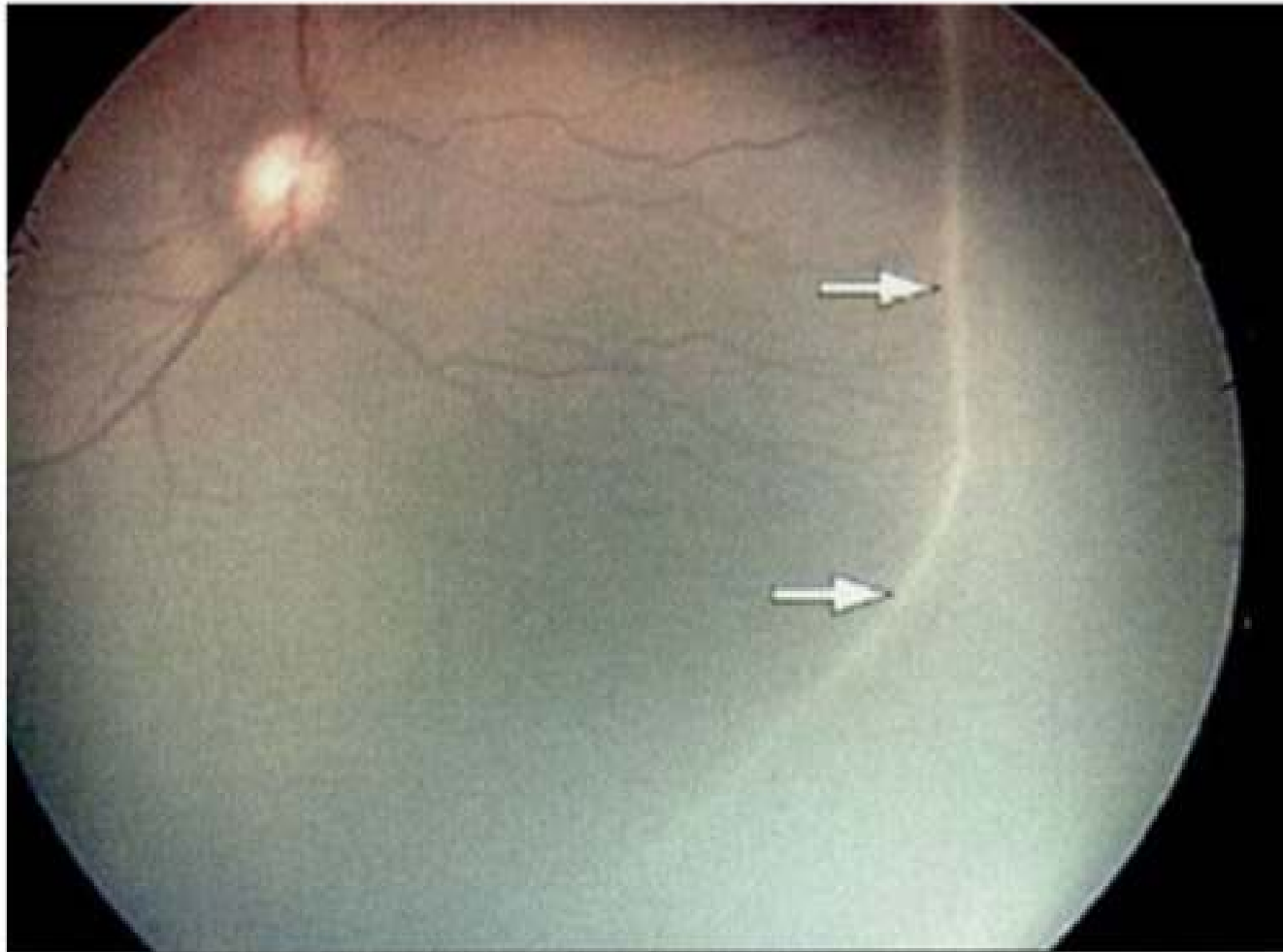
- Disease of retinal vasculature in immature retina of a premature neonate.
- **Results from** interruption of normal vascularization.
- **Characterized** by vaso-obliteration/ vaso cessation followed by abnormal neovascularization and ultimately cicatrisation.
- **Risk factor :**
 - ✓ Birth before 32 weeks' of gestation.
 - ✓ Birth weight of less than 1500 g.
 - ✓ **supplemental oxygen, hypoxemia.**



Stages (1-5)

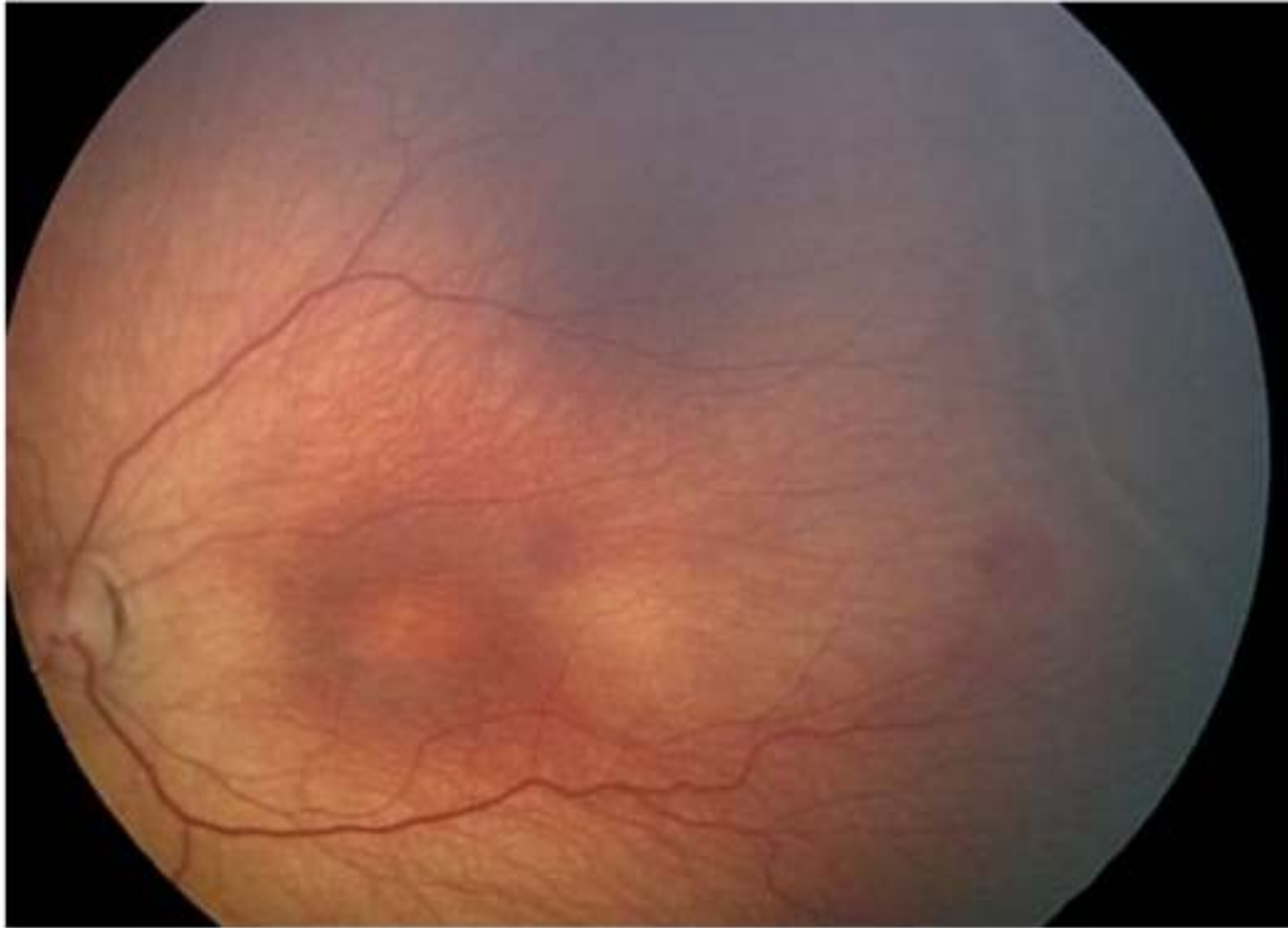
Stage 1:

Demarcation line -a flat, thin, whitish, clear-cut demarcation between vascularised and avascular retina



Stage 2:

Elevated ridge - demarcation line has "3D" appearance & extends anteriorly from the retinal plane as a ridge into the vitreous



Stage 3:

Neovascularisation - Extraretinal fibrovascular tissue begins to grow on the top of the ridge or posterior to the ridge and extends into the vitreous.

“Pop-corn lesion”

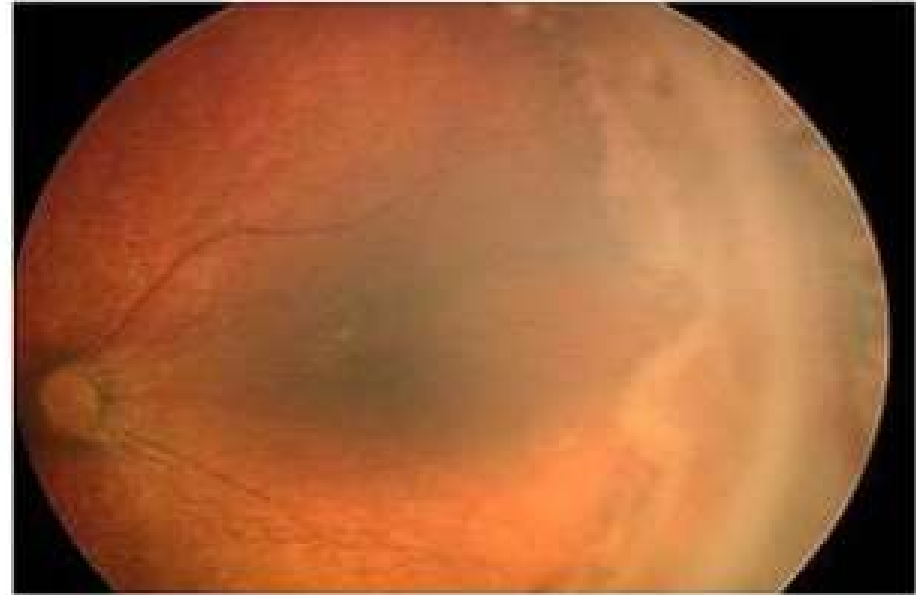


Stage 3 retinopathy of prematurity In stage 3 ROP, new blood vessels and fibrous tissue grow along the ridge and extend into the vitreous.

STAGE 4 (Partial RD)

4A: Extra foveal

4B: Foveal

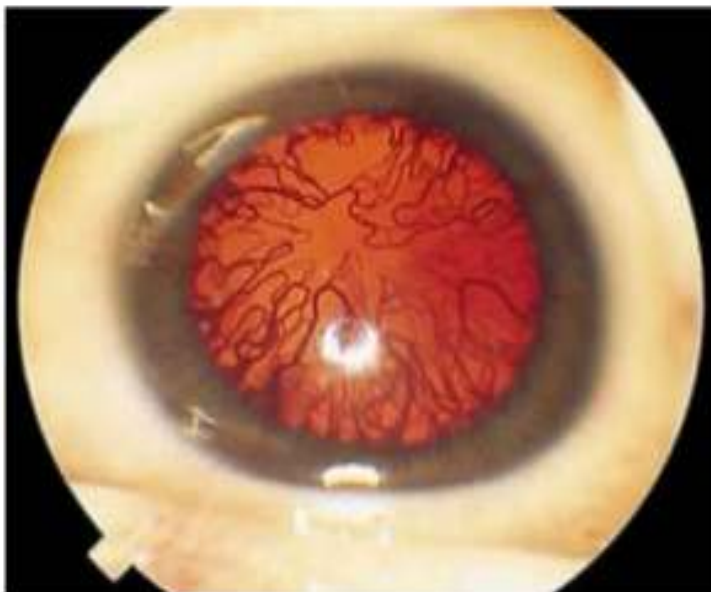


STAGE 5 (TOTAL RD)



Plus Disease

- When the blood vessels of the retina have become enlarged and twisted, **indicating a worsening of the disease.**
- poor pupil dilatation.
- vitreous haze.
- vascular engorgement of the iris with extension onto anterior lens surface.



TREATMENT

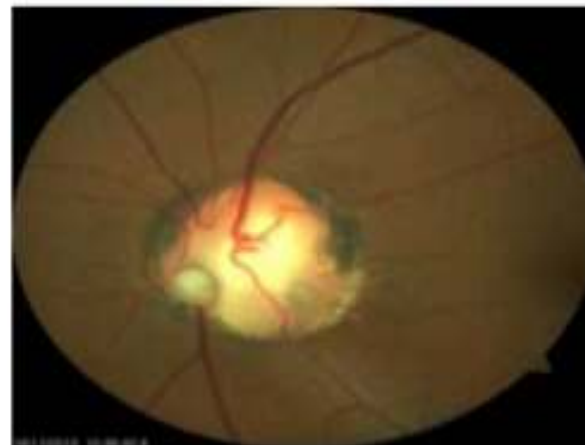
- RETINAL ABLATION
 - CRYO
 - LASER
- SCLERAL BUCKLING
- VITRECTOMY
 - LENS SPARING
 - With LENSECTOMY

Coloboma

- **Congenital coloboma** is developmental defects.



- either **retinal coloboma**
- Or **optic nerve coloboma** can cause leukocoria.



- Other optic disc abnormalities such as a **(morning glory disc)** myelinated nerve fibers.





THE

TAKE-HOME MESSAGE

THANKS

